



CASE REPORT

Adult type rhabdomyoma in a child

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KEYWORDS

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Summary Adult rhabdomyoma (AR) is a rare benign tumor of the skeletal muscle that usually occurs in the head and neck region. AR present in a mean age of 50 years and occur commonly in males than females (4:1). Treatment is excision and recurrence is rare. The aim of this paper is to describe the clinicopathologic features of an AR and its differential diagnosis to avoid unnecessary aggressive treatment and present the fifth AR occurred in a child.

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Introduction

Rhabdomyomas are rare benign mesenchymal tumors composed of striated muscle cells and distinguished in cardiac and extracardiac localisations.^{1–5} Extracardiac type may be classified as genital, fetal and adult type. The genital type rhabdomyomas are slow growing lesions of the vagina or vulva of young middle-aged women. Fetal type is the microscopic variant of adult rhabdomyoma (AR). Although the neoplastic cells are elongated and less differentiated in the fetal type, the neoplastic cells closely resemble to normal muscle in the adult type. Fetal type may be confused with rhabdomyosarcoma.^{1,2,5,7} AR is an asymptomatic, well-defined submucosal solitary mass, although it may be multifocal and is deeply located.^{1–3,6–8} The most frequently reported oral sites for AR are floor of the mouth, soft palate, tongue and buccal mucosa. Treatment of the

lesion is complete excision and recurrence is rare. AR present at a mean age of 50 years and occur more commonly in males than females (4:1).^{2,9} There are only four adult type rhabdomyoma cases previously reported in children.^{6,10–12}

The aim of this paper is to describe the clinicopathologic features of an AR located in the dorsolateral part of the tongue and call attention to this uncommon tumor in the differential diagnosis to avoid unnecessary aggressive treatment and present the fifth adult rhabdomyoma occurred in a child.

Case report

An eight year-old child was presented to the clinic with a complaint of an asymptomatic swelling on the left posterior part of the tongue. It was a slowly growing mass and was noticed 1 month prior to the clinic visit. Her speech and swallowing was slightly affected. However, no difficulty with breathing was noted. She had neither systemic disease and current medication nor hereditary disease in familial history.

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Figure 1 Pedunculated, non-tender mass similar in colour to the surrounding tongue mucosa.

Intraoral examination was revealed a pedunculated and non-tender mass on the left dorsolateral part of the tongue (Fig. 1). It was slightly firm and fixed at palpation. There was no pain and history of paresthesia. Surface of the lesion was similar to the colour of the surrounding tissues. Extraoral examination revealed no abnormalities. There was no palpable lymphadenopathy.

The clinical features was similar to a benign lesion, therefore treated with conservative simple excision and the lesion was excised from the surrounding musculature of the tongue. The biopsy site was primarily closed with 3.0 silk sutures.

Histopathologic examination was performed by Oral Pathology Department of Gazi University Dental Faculty. Macroscopically, the specimen was a white, smooth, firm nodule measuring $1,0 \times 1,0 \times 0,8$ cm. Multiple sections of

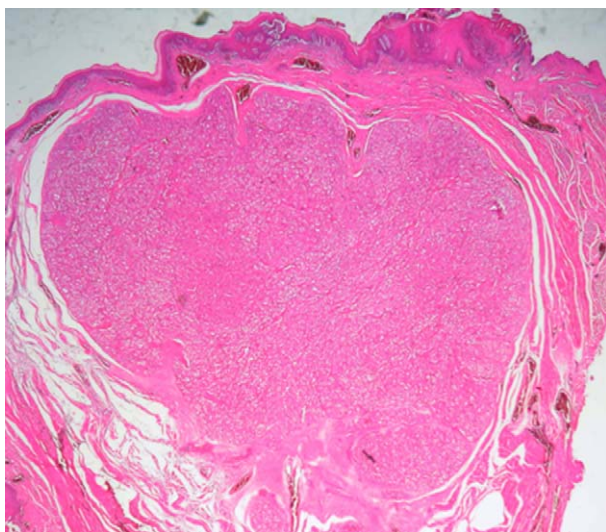


Figure 2 Well circumscribed nodular tumor below mature squamous epithelial (hematoxylin–eosin $\times 20$).

the tumour were prepared routinely and stained with hematoxylin and eosin, periodic acid-schiff (PAS) with and without diastase and phosphotungstic acid hematoxylin (PTAH). For immunohistochemical analysis, sections were reacted with antibodies including muscle-specific actin (+), desmin (+), S-100 protein (–), myoglobin (+).

Microscopically, the tumour was well circumscribed and multilobulated (Fig. 2). The tumour was composed of closely packed, large, round to polygonal cells with finely granular cytoplasm and eccentric vesicular nucleus. Many cells exhibited complete or partial vacuolization of the cytoplasm with occasionally "spider web" configurations. No mitoses were observed (Figs. 3 and 4). Cytoplasmic cross-striations were difficult to identify on routine hematoxylin and eosin stained sections.

But with PTAH stain; both cytoplasmic cross-striations and rod-like crystalloid structures, which stain intensely purple could be identified (Fig. 5). Although the pattern of reactivity varied, tumour cells showed positivity with the muscle-specific actin, desmin and myoglobin antibodies (Fig. 6).

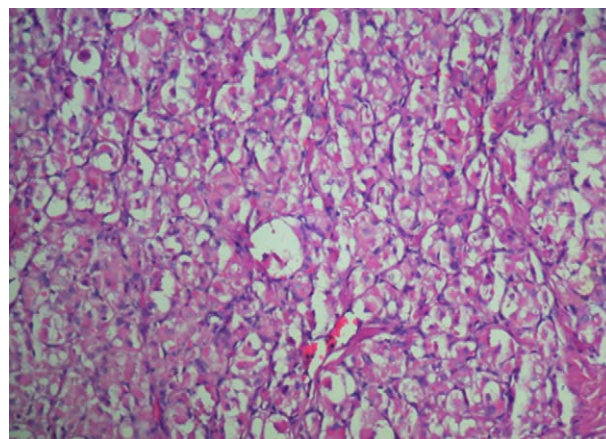


Figure 3 Vacuolated tumor cells involving granules in their cytoplasm (hematoxylin–eosin $\times 200$).

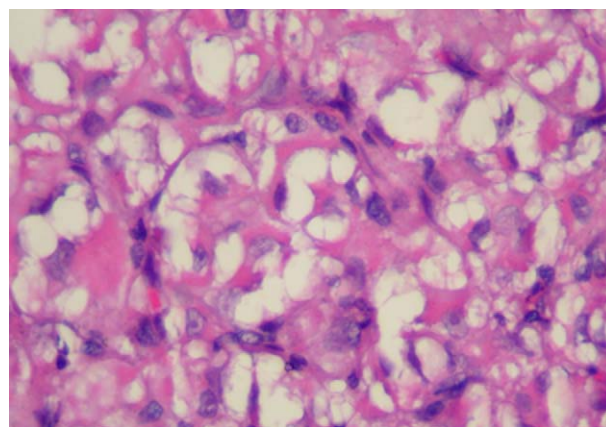


Figure 4 Typical spider cell configuration special to rhabdomyoma (hematoxylin–eosin $\times 600$).

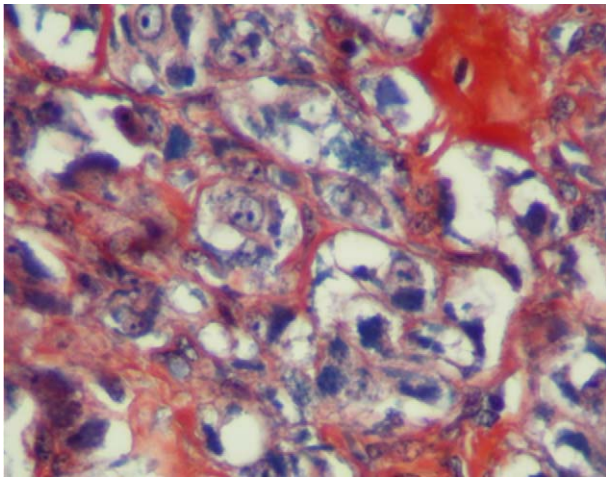


Figure 5 Intracytoplasmic crystalloid granular structures (PTAH $\times 600$).

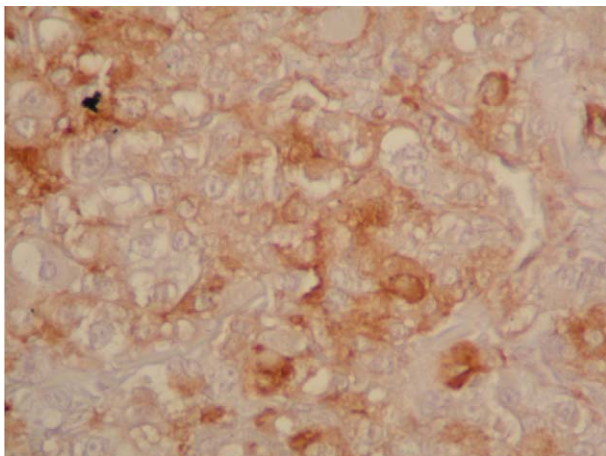


Figure 6 Actin positive tumor cells (ABC $\times 200$).

Discussion

AR is a rare neoplasm of mesenchymal origin representing approximately 2% of all tumors with skeletal muscle differentiation.^{1,13} To date only about 160 cases of the extracardiac type have been reported in the literature.⁸

AR may present with symptoms causing obstruction or remaining asymptomatic for many years.³ Because of a slow rate of growth, they often become quite large before producing symptoms. Among all the cases reported, only 14 was multifocal.^{3,8} Incidence of recurrences reported were 16% and are usually attributed to incomplete initial excision.^{7,13,14} To date, there has been no report of dedifferentiation of a recurrent adult rhabdomyoma to a malignant variety.¹³

Magnetic resonance imaging (MRI)¹⁵ and computed tomography (CT)¹³ findings useful for diagnosis of AR. Moreover, fine needle aspiration biopsy can be performed for the correct initial diagnosis to eliminate aggressive surgery.⁹

The characteristic histologic features of AR are polygonal or ovoid cells with eosinophilic cytoplasm which demonstrates peripheral vacuolization resulting in a "spider web"

appearance and focal cells with cross-striations. The differential diagnosis of AR includes granular cell tumor (Abrikossoff's tumour), hibernoma and rhabdomyosarcomas.^{5,7,14}

Granular cell tumor occurs in the same location as the adult rhabdomyoma and has similar histology. But the cells of granular cell tumour lack the cross-striations and the intracellular vacuolisation. Another different point is typical immunoreactivity for S-100 protein but not for myogenic markers, such as actin, desmin, and myoglobin.¹⁴

The rhabdomyosarcomas and other sarcomas must be distinguished from the AR especially in young patients as an rapidly growing mass, usually associated with pain and paresthesia. The sarcoma cells are less differentiated and they display cytoplasmic atypia and mitosis. Rhabdomyosarcomas are composed of spindle or rounded cells, showing very occasional cross-striations, if any, and atypical and pleomorphic nuclei.^{7,9,13,14}

Hibernoma is another similar lesion, composed of brown fat cells. The cells of hibernoma are smaller, never have cross-striations but contain lipofusion pigment.^{7,14} Sometimes, rhabdomyomas localized in the floor of the mouth may mimic salivary gland tumors, such as oncocytoma or multifocal oncocytosis.¹ Nevertheless, the absence of salivary gland ducts and acini within the lesion and the absence of nucleoli can allow the correct diagnosis, which could be facilitated by the immunohistochemical expression of myogenic markers in rhabdomyoma.^{7,9,13,14}

In the histopathologic examination of our case "spider web" configuration revealed with hematoxylin–eosin and PTAH stain was performed to identify both cytoplasmic cross-striations and rod-like crystalloid structures. Muscle-specific actin, desmin, myoglobin, S-100 were used for differential diagnosis of granular cell tumor.

Although adult rhabdomyoma in a child is an extremely rare pathology, it should be included in the differential diagnosis of the tongue masses of children.

References

1. Favia G, Lo Muzio L, Serpico R, Maiorano E. Rhabdomyoma of the head and neck: Clinicopathologic features of two cases. *Head and Neck* 2003;**25**:700–4.
2. Roberts F, Kirk AJB, More IAR, Butler J, Reid RP. Oesophageal rhabdomyoma. *J Clin Pathol* 2000;**53**:554–7.
3. Liess BD, Zitsch 3rd RP, Lane R, Bickel JT. Multifocal adult rhabdomyoma: a case report and literature review. *Am J Otolaryngol* 2005;**26**:214–7.
4. Rognini RE M, Bruschini L, Leopardi G, Conti S. Adult rhabdomyoma of submandibular region. A case report. *Yl Valsalva* 2002;**78**:172–5.
5. Neville BW, Damm DD, Allen CM, Bouquot JE. *Oral and maxillofacial pathology*. second ed. Philadelphia: WB Saunders; 1995. p. 304–5.
6. Pai GK, Pai PK, Kamath SM. Adult rhabdomyoma of the esophagus. *J Pediatr Surg* 1987;**22**:991–2.
7. Rosai J. *Ackerman's surgical pathology*. eighth ed. Mosby Company; 1995., pp. 2077–8.
8. Delides A. Multifocal adult rhabdomyoma of the head and neck: A case report and the review of the literature. *Eur Arch Otorhinolaryngol* 2005;**262**:504–6.
9. McGregor DK, Krishnan B, Green L. Fine needle aspiration of adult rhabdomyoma: A case report with the review of the literature. *Diagn Cytopathol* 2003;**28**:92–5.

10. Solomon MP, Tolete-Velcek F. Lingual rhabdomyoma (adult variant) in a child. *J Pediatr Surg* 1979;14:91–4.
11. Prisiuda Ala, Sergeev VN, Sarnatskii VM. Rhabdomyoma of the tongue in a 5-year-old child. *Klin Khir* 1976:65.
12. Cacciari A, Predieri B, Mordenti M, et al. Rhabdomyoma of a rare type in a child: case report and literature review. *Eur J Pediatr Surg* 2001;11:66–8.
13. Vermeersch H, van Vugt P, Lemmerling M, Moerman M, De Potter C. Bilateral recurrent adult rhabdomyomas of the pharyngeal wall. *Eur Arch Otorhinolaryngol* 2000;257:24–6.
14. Enzinger FM, Weiss SW. *Soft tissue tumors*. third ed. The CV Mosby Company; 1994., pp. 433–47.
15. Fukuda Y, Okamura HO, Nemoto T, Kishimoto S. Rhabdomyoma of the base of the tongue. *J Laryngol Otol* 2003;117:503–7.